

Supplementary data

Supplementary figure 1: Characteristics of previously reported patients with ECD and sclerosing mesenteritis.

Author (year)	Nb of patient	Gender	Age	Main symptom	Abdominal symptoms	ECD location	Tissue disclosing ECD	BRAF ^{V600E}	Mutation detected	Treatment	Outcome (at publication)
Tan (1995)	1	F	59	Abdominal swelling	Abdominal swelling	Bones, kidney, heart, mesentery, breast	Breast, pericardium	NA	NA	Cyclophosphamide/vincristine/prednisone	NA
Kang (2003)	1	M	53	Dyspnea	No	Bones, heart, lung, mesentery	Pleura	NA	NA	NA	Dead
Perlat (2009)	1	M	31	Bone pain	No	Bones, skin, mesentery	Skin, mesentery	NA	NA	Prednisone/Cladribine Interferon	Died from infection
Lamboley (2010)	1	M	55	Abdominal pain	Abdominal pain	Bones, kidney, vascular, mesentery	Mesentery	NA	NA	NA	NA
Pan (2011)	1	M	69	Vomiting	Vomiting	Bone, kidney, vascular, lung, mesentery	Liver, omentum mesentery	NA	NA	Prednisone	Died of respiratory failure
Sedrak (2011)	1	M	60	Ascites	Ascites	Skull base, mesentery, kidney	Bone marrow	NA	NA	NA	NA
Haroche (2012)	1 (mixed LCH)	W	59	Bone pain	No	Bones, kidney, heart, vascular, orbit, mesentery	Mesentery	BRAF	none	Interferon-alpha Vemurafenib	Alive with disease
Gilortega (2013)	1	M	45	Diarrhea	Diarrhea	Bones, kidney, mesentery	Tibia	NA	NA	Interferon-alpha	Alive with disease
Emile (2014)	1	M	54	NA	NA	Mesentery	Mesentery	W-t	None	NA	Alive with disease
Gianfreda (2015)	4	#1: M #2: F #3: F #4: M	#1:68 #2: 38 #3: 42 #4: 46	#1: Constitutional signs #2: Skins #3: Bone pain #4: Pericarditis	#1: pain/vomiting #2: no #3: ascites #4: no	#1: Bones, kidney, vascular, mesentery #2: Bones, vascular, mesentery, pituitary, CNS, skins #3: Bones, cardiac, vascular, mesentery, endocrine system, lung, lymph node #4: Bones, cardiac, vascular, kidney, mesentery, orbit, endocrine system	#1: NA #2: Skin #3: Kidney and node #4: Kidney	#1: w-t #2: w-t #3: BRAF #4: w-t	#1: NA #2: none #3: none #4: none	#1: Sirolimus/prednisone #2: Sirolimus/prednisone #3: Sirolimus/prednisone Vemurafenib #4: Sirolimus/prednisone	#1: Alive with disease #2: Alive with disease #3: Alive with disease #4: Alive with disease
Franconieri (2016)	1	F	76	Constitutional signs	NA	Bones, kidney, mesentery, vascular, orbital, pituitary, pleural, pericardial	Retro-orbital	W-t	NA	Anakinra Interferon-alpha Canakinumab Vemurafenib	Alive with disease
Goyal (2017)	1	M	76	NA	NA	Bone, bone marrow, CNS, orbit, sinus, kidney, mesentery	NA	NA	NA	Prednisone Anakinra	Alive with disease
Franconieri (2018)	2	#1:M #2:M	#1:68 #2:52	#1: Constitutional signs #2: Constitutional signs	#1: yes #2: yes	#1: Bone, kidney, cardiac, pleural, mesentery #2: Bone, kidney, cardiac, pleural, mesentery	#1: kidney, mesentery #2: kidney, mesentery	#1: w-t #2: BRAF	#1: NRAS	#1: Interferon-alpha Anakinra #2: Steroids-vinblastine Anakinra	#1: Alive with disease #2: Alive with disease
Nikpanah (2018)	4	NA	NA	NA	NA	NA	Mesentery in one	NA	NA	NA	NA

w-t: wild type, CNS: central nervous system; NA: non-available